



# Degeneration of the Frontotemporal Lobar Region and Frontotemporal Dementia

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## DESCRIPTION

The characteristic of the frontotemporal dementia family of illnesses is the development of degenerative disease in localised regions of the cerebral cortex (also termed frontotemporal lobar degeneration). Since Arnold Pick's seminal case report from 1892, cases of elderly individuals with increasing verbal degradation have been documented. His case study, "On the Relationship between Aphasia and Senile Atrophy of the Brain," is still used as a standard for seemingly focal brain symptoms in diffuse or generalised degenerative brain disorders. Simply gradual brain shrinkage, according to Pick, "may provide local disruption symptoms by local accentuation of the diffuse process."

Two concurrent streams of information about focal brain degenerations accumulated in the 1980s and 1990s. Mesulam described six cases of progressive aphasia in 1982. These patients did not advance to more extensive dementia. Instead, their condition deteriorated over time. Numerous other cases have been reported after Mesulam's publication, and Mesulam's team has added more reviews. Primary progressive aphasia (PPA), the name of this condition, is now recognised as a syndrome.

The PPA syndrome was later identified as a condition characterised by progressive aphasia over a two-year period without general cognitive decline or dementia. According to Kirshner et al., many individuals experience more generalised dementia later on in the course of the illness. Less frequently, reports have been made of isolated right frontal or temporal degeneration. Failure to recognise family members (prosopagnosia), forgetting topographic relationships, and other similar issues affect these patients.

Frontal lobe dementia instances with progressive frontal lobe dysfunction have been reported in England and throughout Europe. Neary and Snowden described a syndrome with initial symptoms that were suggestive of psychiatric disorder in a number of case reports. However, with time, the following frontal lobe behavioural anomalies developed:

- Disinhibition
- Impulsivity
- Impersistence
- Inertia
- Social awareness decline
- A disregard for personal hygiene
- Rigidity of the mind, stereotypical conduct

Utilization behaviour, or the propensity to pick up and handle any nearby thing language disorders like diminished speech output, mutism, echolalia, and persistence were included in these descriptions.

Frontotemporal Lobe Dementia (FTD), also known as frontotemporal lobar degeneration, is the unified name for the conditions known as frontal dementia in Europe and primary progressive aphasia in North America (FTLD). The terms behavioural variant frontotemporal lobe dementia (bvFTD) and frontal variant frontotemporal lobe dementia are interchangeable terms for the frontal lobe condition identified by Neary and Snowden (fvFTD). Progressive nonfluent aphasia, semantic dementia, and logopenic progressive aphasia are the three groups of progressive aphasias.

Frontotemporal dementia is now a catch-all phrase used to describe various clinical syndromes of frontal dementia or progressive aphasia. Frontotemporal lobar degeneration is another term for the disorders connected to frontotemporal lobe dementia syndromes. The two phrases are essentially used interchangeably throughout this evaluation.

## TREATMENT

The majority of current studies have focused on identifying FTD and comprehending its pathophysiology. Specific medical treatments might develop as we gain a better understanding of the aberrant gene products. However, there are currently very few medical treatments available.

For the patient, carers, and family members, social interventions, counselling, and speech-language-cognitive therapy to enable the

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use of spared functions may make the condition easier to endure. It is yet to be established whether behavioural therapies decrease the disease's course.

While trazodone and selective serotonin reuptake inhibitor (SSRI) antidepressants are frequently suggested, all current pharmaceutical therapies are unproven. There is no proof that FTD involves a cholinergic deficit, and there is no clinical evidence of benefit, despite the fact that cholinesterase inhibitors, licenced for Alzheimer's disease, are occasionally used in this illness. According to anecdotal evidence, some patients' memories may become better while others' behaviours appear to

get worse. Similarly, memantine (Namenda), a medication, has been used to treat FTD, but two recent modest clinical trials failed to show any improvement. Future discoveries regarding the molecular biology and genetics of these illnesses are anticipated to pave the way for disease-modifying therapies.

## PATIENT EDUCATION

Visit the Brain and Nervous System Center, Dementia Overview, and Dementia Medication Overview for more on patient education.